

Strategic Plan
National Institute on Deafness and Other Communication Disorders (NIDCD)
FY 2006-2008

OVERVIEW

We depend on our ability to communicate effectively to function in our modern society. Approximately one of every six Americans experiences some form of communication disorder, which, for the purposes of the National Institute on Deafness and Other Communication Disorders (NIDCD) includes disorders that affect hearing, balance, smell, taste, voice, speech, or language. While science and technology have increased our capacity and need for communication, many aspects of contemporary life remain profoundly difficult for individuals with communication disorders. These disorders often compromise social, recreational, emotional, educational, and vocational aspects of an individual's life. The cost of these disorders in quality of life and unfulfilled potential is substantial. As the population ages and as survival improves for medically fragile infants and individuals who have sustained injury or acquired disease, the number of citizens with communication disorders will continue to increase.

What Is the NIDCD?

In 1988, Congress established the NIDCD as a separate institute within the National Institutes of Health (NIH). The mission of the NIDCD is to conduct and support research and research training in the normal and disordered processes of hearing, balance, smell, taste, voice, speech, and language. These processes of sensing, interpreting, and responding are fundamental to the way we perceive the world and to our ability to communicate effectively and efficiently. Basic and clinical research focused on understanding the normal processes and disorders of human communication is motivated by intrinsic scientific interest, by the goal of allowing more individuals to reach their potential, and by reducing the cost, both tangible and intangible, to individuals and to the nation.

The NIDCD Strategic Plan

Historically, the NIDCD developed a strategic plan for human communication research, segments of which were updated by panels of extramural scientific experts from one or two of its seven mission areas on a regular cycle. In 1999, the NIDCD embarked on a new strategic planning process by convening a group of distinguished scientists, clinicians, and members of the public to provide recommendations for a Strategic Plan for the NIDCD for Fiscal Years 2000-2002. The NIDCD charged the Strategic Planning working group with identifying areas of research within the NIDCD mission, and to consider the research currently supported by the Institute, as well as NIH-wide scientific initiatives. In addition, individuals and representatives of public and private organizations as well as scientific organizations provided input to the Strategic Planning Working Group. These responses helped ensure that the public's perspective was assimilated into the recommendations for the Strategic Plan.

Following a review of this input, the Strategic Planning Working Group defined broad scientific areas for NIDCD-supported research over a three-year period. The final draft of the plan was

discussed in detail at the National Deafness and Other Communication Disorders (NDCD) Advisory Council in May 1999, and implemented shortly thereafter. The plan was reviewed and updated by the NDCD Advisory Council in 2002 for Fiscal Years 2003-2005 and in 2005 for Fiscal Years 2006-2008.

Below are highlights of the compelling needs of individuals who have communication disorders and the extraordinary research opportunities in the NIDCD's mission areas that address these needs.

BACKGROUND

Diseases and Disorders of Human Communication are Significant Health Problems

Statistics

Birth and Early Childhood

- ♦ Approximately 2-3 out of 1,000 babies are born each year in the United States with a detectable hearing loss, which can affect their speech, language, social, and cognitive development.
- ♦ About 8 percent of American children in kindergarten have a disorder called specific language impairment (SLI). These children have difficulty developing and using language. These difficulties affect not only speaking but also reading and writing tasks.
- ♦ Middle ear infections (otitis media) are the most frequent reason that a sick child goes to the doctor. The estimated total cost of otitis media in the United States is \$5 billion per year. Children with otitis media can suffer temporary hearing loss during the infection, often for some time during treatment, but may suffer permanent hearing loss.
- ♦ Approximately 1 out of every 200 American children is diagnosed with autism, a disease that interferes with normal language and social development. Boys are four times more likely to be born with autism than girls. Girls with the disorder, however, tend to have more severe symptoms and greater cognitive impairment.
- ♦ Approximately 1 million American children stutter. Stuttering affects individuals of all ages, but occurs most often in young children who are beginning to develop language skills. Boys are three times more likely to stutter than girls.
- ♦ Approximately 5 percent of American children entering first grade have noticeable speech (phonological) disorders, ranging from a few substituted and missing sounds to serious impairments that make their speech difficult to understand. These speech disorders are about 1.5 times more prevalent in boys than girls. The majority of these speech disorders have no known cause.

- ♦ Flavor is the primary determinant of whether food is accepted by children under the age of two. About a quarter of American infants and toddlers between the ages of 7 and 24 months consume no discrete servings of vegetables and about a quarter consume no fruits on a given day, which has important nutritional consequences.

Adulthood

- ♦ Approximately 15 percent (or 32.5 million) of American adults report some degree of hearing difficulty.
- ♦ There is a strong relationship between age and reported hearing difficulty: 18 percent of American adults aged 45-64 years, 30 percent of adults aged 65-74 years, and 47 percent of adults aged 75 years or older. The age-adjusted prevalence of reported hearing difficulty is higher for men (18.6 percent) than for women (12.6 percent).
- ♦ Approximately 10 percent (or 22 million) of American adults between 20-69 years old have suffered permanent damage to their hearing from exposure to loud sounds or noise at work or in leisure time activities. Noise-induced hearing loss is more prevalent in men than in women.
- ♦ Nearly 1 million American adults have aphasia (a language disorder) resulting from a stroke or other brain injury.
- ♦ More than 6 million adults over the age of 60 have swallowing problems. Many individuals with swallowing disorders, e.g. resulting from stroke, are at risk for aspiration pneumonia.
- ♦ 55,000 Americans each year develop cancer of the head and neck. Treatment for these cancers and other types of cancer may subsequently result in a loss of hearing, balance, and the ability to speak and to swallow.
- ♦ Approximately 4 percent (or almost 8 million) of American adults report a chronic problem (lasting 3 months or longer) with balance, while an additional 1.1 percent (or 2.4 million) of American adults report a chronic problem with dizziness alone. Overall, it is estimated that the cost of medical care for patients with balance disorders exceeds \$1 billion per year in the United States.
- ♦ Balance disorders are a major cause of falls by American elderly, and are the most common reason individuals over the age of 75 visit their primary care physician. Patient care costs for these falls are more than \$8 billion per year.
- ♦ An estimated 24.5 percent (or approximately 15 million) of Americans aged 55 years or older suffer olfactory impairment, which increases with age. Approximately 30 percent of Americans between the ages of 70 and 80 and 62.5 percent over age 80 experience problems with their sense of smell. Impairment in olfaction can have serious

consequences, such as the inability to detect the foul smelling odorants that are added to natural gas as a warning sign of leaks.

What Research Progress Has Been Made?

What Is Known? Research has produced many important discoveries and many new technologies to identify and help individuals with communication problems. As a result of research:

- ♦ Vaccines now prevent many illnesses such as measles, mumps, meningitis, and rubella that once were major causes of hearing loss.
- ♦ Understanding the consequences of reduced hearing in childhood has led to prompt treatment of middle ear infections in children, preventing hearing loss later in life and helping children to thrive.
- ♦ Genetic mutations that lead to inherited forms of hearing loss have been identified and the function of many of the proteins they encode has been elucidated. The era of precise genotype-based diagnosis is at hand.
- ♦ Many genes that control the development of the inner ear and the organization and orientation of sensory hair cells within it have been identified.
- ♦ Molecules that are critical to the function of the sensory hair cells have been identified. There has been significant progress in defining how they are organized and how they function in the hearing process.
- ♦ Newborn babies with hearing loss and toddlers with language problems are identified at an early age so that developmental consequences are minimized through prompt and early involvement in intervention.
- ♦ Understanding the ototoxic action of antibiotics has led to the development of strategies for minimizing damage to hearing.
- ♦ Understanding that exposure to noise results in hearing loss has led to the increased use of ear protection.
- ♦ More is known about how infants who are deaf learn sign language and how sign language compares with spoken language.
- ♦ The amount of linguistic input that children receive affects the rate at which they learn language and perhaps their eventual language proficiency.
- ♦ More is known about reading ability in adults who are deaf and this information may lead to improved methods of reading instruction.

- ◆ Knowledge of the biology of neurons continues to grow. Synapses, the points of contact between neurons, have been shown to be highly plastic and are capable of change at all ages. For example, this is important for understanding age-related hearing loss as well as the restoration of hearing through cochlear implants.
- ◆ Cochlear implants allow children who have a profound hearing impairment to attend mainstream schools and allow adults to communicate more effectively.
- ◆ Hearing aids are designed to work better in noisy environments, for example, by detecting the direction from which sounds arise.
- ◆ It has recently been discovered that short electrodes for cochlear implants can be used together with hearing aids for some common age-related hearing disorders.
- ◆ Advances in technology and science have created new opportunities to design devices that restore or improve function for individuals with balance, voice, and speech disorders. For example, improvements have been made in vestibular devices to assist individuals with a balance disorder, electronic larynxes (the organ that produces voice), and computer-aided speech devices.
- ◆ The genes for olfactory receptors have been identified. This pioneering advancement, which earned the discoverers the 2004 Nobel Prize in Physiology or Medicine, together with the discovery of the genes for many taste (gustatory) receptors, open new frontiers in understanding chemosensory disorders.
- ◆ Genetic studies have begun to identify the genes that contribute to stuttering, phonological disorders, and language disorders.
- ◆ New surgical approaches and combinations of chemotherapy and radiation have improved rates of preservation of voice and speech when treating cancers of the head and neck.

What Needs to Be Studied? Despite the considerable advances that have been developed to help individuals with communication challenges, more work remains to be done and questions remain unanswered. For example, more studies are needed to help answer the following general questions:

- ◆ How does sensory processing function in complex, real-world situations, that challenge the nervous system well beyond the relatively simple, unimodal stimuli typically used to analyze communication disorders in the laboratory?
- ◆ How and where in the nervous system are certain communication disorders generated, such as tinnitus (ringing in the ears or hearing phantom sounds), so that treatments can be optimally targeted?

- ♦ Given our new understanding of the genetic and molecular basis of many communication disorders, is it possible to use genetic or molecular interventions to address these disorders?
- ♦ How can existing or new diagnostic tools, such as brain imaging, gene-based diagnostics, and computer-based testing programs help doctors choose the best treatment for individuals with communication disorders?
- ♦ How do communication patterns change during the natural history of a disease, particularly in aging or neurodegenerative diseases, and how can this information be used to deliver the most effective treatments?
- ♦ Why does a particular treatment for a communication disorder work well for some individuals but not for others?
- ♦ What methods can be used most efficiently to identify infants at risk for communication disorders and what types of intervention would be most beneficial?
- ♦ What are the best ways to help children with communication disorders learn to communicate and develop learning skills?
- ♦ How can devices and treatments for communication disorders be improved or developed? What techniques optimally assess performance so that these devices can be adjusted optimally and so that improvement can be measured objectively? Which of these devices or treatments can help the largest group of people who have a particular disorder?
- ♦ How can basic and clinical research and research training be used to address the need to eliminate gender, racial/ethnic, or socioeconomic health disparities in communication disorders?

RESEARCH AREAS THAT OFFER EXTRAORDINARY SCIENTIFIC OPPORTUNITY

With help from scientists and the public, the NIDCD has identified four significant research areas that offer extraordinary research opportunities in the field of human communication sciences. These areas are:

- I. Determine the Molecular and Epidemiological Bases of Normal and Disordered Communication Processes
- II. Study the Development, Deterioration, Regeneration, and Plasticity of Processes Mediating Communication
- III. Study Perceptual, Cognitive, and Sensorimotor Processing in Normal and Disordered Communication

IV. Develop and Improve Devices, Pharmacologic Agents, and Strategies for Habilitation, Rehabilitation, and Prevention of Human Communication Disorders

Where Do Future Research Opportunities Exist?

These four research areas are described in greater depth in the following section. A detailed list of the NIDCD's research priorities is included for each area.

I. Determine the Molecular and Epidemiological Bases of Normal and Disordered Communication Processes

Scientists know that genes play a key role in many communication diseases and disorders; however, this area of research requires much more intense study and more research is needed on non-genetic factors that also affect communication processes, such as infection, toxins, and environmental exposures. Both of these areas (molecular and non-genetic) are research priorities for the NIDCD.

Understanding Molecular Causes (Proteomic and Genomic)

One of the most rapidly developing areas of research involves determining the identity, structure, and function of genes, a discipline referred to as structural and functional genomics. The Human Genome Project has shown that human beings have about 22,000 different genes. Considerable progress has been made in identifying which genes are involved in human communication and how they are altered in individuals with communication disorders. Hereditary disorders are associated not only with abnormalities in single genes but from combinations of particular forms of genes. In fact, genetic mutations are estimated to cause or play a role in a wide variety of communication disorders, including at least one-half of all cases of congenital or childhood-onset hearing loss. Individual variations in the severity of hearing loss are common and typically attributed to environmental factors and modifier genes. Modifier genes in the genetic background of an individual can affect the severity of communication disorders caused by a mutation. Understanding the genetic basis of hereditary disorders can inform clinicians which treatments are effective and allow families to make informed decisions as they deal with those disorders. Much of the success and progress in genomics is a direct result of the willingness and generosity of families with hereditary communication disorders who agree to participate in studies with clinicians and scientists. Clearly, without them, research in this field would not have advanced to its current state of knowledge.

Genes influence cells by determining whether certain proteins are made and, if they are made, by influencing their structure and function. Proteomics is the study of how proteins interact within cells. Proteins are the building blocks of all living cells. They carry out the tasks that allow cells to grow and divide in development, and sustain mature cells. The cells, in turn, form every internal system in the human body. Mutations in one gene can have a dramatic effect on complex functions such as hearing, balance, smell, taste, voice, speech, and language. Understanding how the function of proteins is altered in individuals with communication

disorders of either genetic or non-genetic origin, is an essential first step in developing precise molecular diagnoses, pharmacological treatments, and behavioral interventions.

Mutations in genes contribute to numerous communication disorders, in some cases directly by causing a critical group of cells to malfunction, and in other cases indirectly by increasing the body's sensitivity to damage from infections, certain drugs or medications, or exposure to loud noise over an extended period. Continued research is needed to identify and characterize genes and modifier genes and to understand their function in complex communication disorders with multiple deficits that overlap with other conditions (such as hearing loss, stuttering, speech sound disorders, autism, and dyslexia). This knowledge will enable more accurate diagnosis and classification of individuals with communication disorders. This knowledge can also be used in the long-term planning of clinical interventions. For example, children diagnosed with a mild hearing loss at birth with a gene mutation that will cause progressive hearing loss and/or deafness by their teen years may receive and benefit from early education programs so that they may achieve their full potential.

Understanding Non-Genetic Causes (Infectious, Environmental, and Toxic)

Not all communication disorders have a genetic basis. Some are rooted in experiential factors, and others result from a combination of genetic and experiential factors. For example, hearing loss can occur as a result of infections, noise damage, or toxicity associated with certain medications or other chemicals. Hearing loss in infants can result in difficulty learning to speak or understanding language later in life, if appropriate education and training are not provided. At any age, impaired language skills affect a person's ability to function in today's complex, communication-driven society. Besides childhood hearing disorders, speech and language impairments can also be caused by an injury to the brain or a problem in brain development. Diseases of the larynx can be caused by infections or by the presence of a tumor. Further, in occupations with high voice usage, such as teaching, voice problems limit the ability to perform certain tasks at work, result in missed workdays, and can sometimes lead individuals to consider changing occupations. More research is needed to identify a multitude of causes and mechanisms that result in communication disorders.

Strategic Plan Research Agenda -- Priority Area I

Research is needed to help determine the molecular (proteomic and genomic) and non-genetic (infectious, environmental, and toxic) bases of communication processes in the following areas:

- ♦ Use genomic, proteomic, informatic, bioinformatic, and expression profiling technologies, as well as other molecular biologic and genetic approaches, to understand the molecular bases of normal and disordered human communication, including gene identification, regulation, expression, and associated mutations.
- ♦ Bring emerging technologies in genetics and molecular biology (including DNA microarrays, biomarker identification, and other genomic strategies) to the clinical setting and encourage multidisciplinary approaches to the prevention, diagnosis, and treatment of communication disorders.

- ♦ Encourage multidisciplinary collaboration between scientists in fields such as chemistry, biology, pharmacology, genetics, medicine, etc., to develop prevention, detection, diagnostic, and treatment methods for communication disorders.
- ♦ Study the common variations, including modifications, in human DNA and their impact on susceptibility to a range of human communication disorders.
- ♦ Investigate complex and multifactorial disorders of human communication caused by the interactions of several different genes. Identify and analyze factors that influence variability and susceptibility to disease and response to treatment.
- ♦ Develop *in vitro* and animal model systems to study the function of specific disease genes, identify and isolate specific cell populations, and investigate cellular processes by using such techniques as gene and protein expression systems, organ and cell culture systems, and stem cell research.
- ♦ Explore the pathogenesis, treatment, and prevention of viral and bacterial infections that may contribute to communication disorders.

II. Study the Development, Deterioration, Regeneration, and Plasticity of Processes Mediating Communication

Understanding how the brain and the sensory organs involved in human communication recover or adapt to injury or damage is another research priority for the NIDCD.

Increasing the Potential for Recovery -- How the Body Creates New Cells

Many parts of the human body that are damaged by illness or injury can heal by regenerating healthy cells to replace damaged or lost ones but other parts of the body, including hair cells of the inner ear, do not regenerate spontaneously. Recovery from injury also results from adaptive changes that allow other parts of the body to resume functions that were lost.

Until recently, scientists believed that the highly specialized hair cells of the inner ear, which are critical for hearing and balance, could never be replaced if they were injured or destroyed. It has recently been shown that mammalian hair cells can be replaced by regenerating healthy hair cells from supporting cells if certain genes are expressed. This finding inspires hope that damaged inner ear hair cells in humans, one of the major causes of hearing loss, can be repaired or replaced. To make it possible to regenerate hair cells in humans safely requires further fundamental studies. Before genes are introduced into humans, it is essential that the molecular machinery that is used to deliver the genes is safe and that the functional consequences of having those genes in the ear over long periods are fully understood.

Human olfactory receptor neurons in the nose show a remarkable ability to regenerate throughout life. We need to study the unique ability of these regenerated cells to make proper

connections to brain regions that recognize and discriminate odors. Translating basic knowledge of how olfactory receptor cells regenerate to clinical uses, including identifying the factors that make regeneration possible, could lead to clinical intervention strategies that promote nerve cell regeneration not only in the nose but also throughout the nervous system.

The central nervous system adapts to changes by reorganizing connections among neurons. Its “plasticity” allows us to learn as well as to cope with injury but these adaptations can also cause problems such as substance abuse. When a part of the brain involved in speech and language is injured by a stroke or an infection, changes in the connections between neurons may allow other parts of the brain to learn and/or take on that function. Understanding how such adaptive changes are limited opens the door for treatments that optimize the ability of the brain to make such changes.

Adaptation to changes can also have deleterious consequences. After damage to the auditory system, many people acquire tinnitus, which can be very disturbing. Most treatments available today involve learning to live with tinnitus or covering up the tinnitus with other sounds. Currently, there is no cure for tinnitus; however, understanding where in the auditory system the changes occur that underlie tinnitus and what the changes are is a necessary first step in developing treatment. Plasticity is also important to the function of the vestibular system. Damage to one vestibular organ is rapidly compensated by changes that occur in the brain. Understanding how the vestibular system adjusts to changes may be an important step in developing treatments for vestibular disorders.

Adults who suffer brain damage from a stroke often experience problems expressing their thoughts through speech and language. These speech and language disorders severely limit their ability to communicate and often restrict vocational/occupational opportunities, which in turn, decreases quality of life. In contrast, infants and young children who have suffered similar brain damage from birth injuries, childhood trauma, or extensive brain surgery are more likely than adults with similar damage to develop or recover speech and language abilities. Additional research is needed to uncover and understand how and why young children can recover from, or adapt to, severe brain damage, so that new methods can be developed to promote recovery in adults.

Early in life, sensory cells in the hearing and balance organs of the inner ear develop connections with specific brain regions. At certain times in a young child’s life, the brain is more able to form these connections. The ability to develop these critical brain connections may be lost forever if these time-sensitive opportunities are missed because sensory information is not being properly transmitted to the brain, for example, in an infant with severe hearing loss that has not been detected. Research is needed to identify these critical “windows of opportunity” for developing brain connections essential to communication. Important research findings in this area have already motivated major United States public health efforts, such as screening millions of newborns for hearing loss each year.

Stem cells have the ability to regenerate and differentiate into specialized cells. If scientists understand the events that allow stem cells to differentiate (become a certain type of tissue), they may be able to use these cells for a variety of therapeutic purposes, driving the regeneration of

cells damaged or destroyed in the sensory organs of the ear, nose, and mouth as well as in the brain.

Strategic Plan Research Agenda -- Priority Area II

Research is needed to determine how development, deterioration, regeneration, and plasticity contribute to the communication process in the following areas:

- ♦ Characterize age-related changes in structural and functional plasticity of communication processes. For example, the inner ear (and other structures) has a complex structure and any variations in its early development may contribute to dysfunction or malformation. Therefore, it is important to understand the normal development of structures/processes related to communication at the molecular level so that gene variants that may cause subsequent malformations can be identified. This information may also lead to further understanding of regeneration/degeneration in these structures.
- ♦ Develop and apply techniques such as functional imaging [e.g., functional magnetic resonance imaging (fMRI) and positron emission tomography (PET)] to assess structural and functional plasticity.
- ♦ Determine the cellular and molecular mechanisms underlying sensory cell degeneration and regeneration (cochlear and vestibular hair cells, olfactory and gustatory cells), which may lead to the development of new therapeutic interventions.
- ♦ Use *in vitro* assays to investigate molecular factors involved in stimulating embryonic and adult stem cells to differentiate into specific cell types used in communication.
- ♦ Use cultured and organ tissue/cell systems to investigate the importance of signals in the microenvironment of specific cell types involved in communication processes.
- ♦ Investigate cellular and molecular protective mechanisms used by the body to protect auditory, vestibular, olfactory, and gustatory receptor cells. Develop methods to enhance these processes to improve survival of these sensory cells following trauma or disease.
- ♦ Determine and categorize mechanisms involved in the development, maturation, aging and recovery of function needed for communication, including cell proliferation, differentiation, neuron axon targeting, pattern formation, cell death, and survival.
- ♦ Understand changes in the brain that result from the loss of sensory input, e.g., deafness, anosmia (inability to smell), or agusia (inability to taste). Understanding changes that result from a loss of sensory input is also important for making the best use of drug treatments, behavioral interventions, and assistive devices.
- ♦ Develop animal models to study the underlying processes of communication disorders. For example, because of the subjective nature of tinnitus, it has been difficult to develop animal models; however, efforts need to continue in spite of these limitations.

- ♦ Determine the optimal clinical interventions by incorporating knowledge from studies of neuroplasticity (as it occurs in normal development and in response to injury) to include information on the timing, quantity, and method of treatment.
- ♦ Investigate the molecular mechanisms of neurotransmission at peripheral and central synapses associated with hearing, balance, smell, taste, voice, speech, and language to understand normal function and function under disease conditions with the objective of treating these disorders with pharmaceuticals.

III. Study Perceptual, Cognitive, and Sensorimotor Processing in Normal and Disordered Communication

Obtaining a more detailed understanding of how the brain acquires, organizes, and interprets information is another research priority for the NIDCD.

Perceptual Processing

Human communication relies on complex perceptual skills by using the senses (hearing, vision, touch, pain, smell, and taste) to receive and interpret information from the outside world. Human communication also requires mental abilities, such as attention and memory. Scientists do not fully understand how all of these processes work and interact, or how they malfunction when there is a communication disorder. They do know that many communication disorders occur even when the peripheral sensory organs appear completely normal.

Recently, new methods have been developed to study what happens after sense organs receive information. With computerized imaging, it is now possible to view directly regions of the brain at work. This advanced technology allows scientists to see changes as information flows from sensory organs to the brain. For example, a functional magnetic resonance imaging (fMRI) scan of the brain can be used to observe activity as language information (written, spoken, or signed words) is received, processed, and interpreted by the brain. Research using brain-imaging techniques is now allowing scientists to challenge the old belief that there is a fixed part of the brain just for organizing language. Studies in both adults and children indicate that brain organization can be modified. After an injury to either the right or left side of the brain, the organization of language that normally takes place there begins to take place in other brain regions. For some individuals, this rerouting may allow relatively normal language abilities to be restored. Scientists cannot obtain detailed information on human speech and language by studying animals, so new imaging studies involving humans are crucial.

Moreover, imaging studies are already giving scientists new insights into auditory processing. Some of these studies are directed at understanding auditory processing disorders (APD). APDs adversely affect learning and language acquisition. Many of the symptoms related to APD are described differently by separate people. Studies of twins have determined that certain auditory processing abilities are highly heritable. It is necessary to continue to evaluate which aspects of

APD are heritable through twin studies and to conduct family studies to estimate the number of genes that may contribute to APD and the magnitude of their effects.

Cognitive Processing

Improved methods of functional brain imaging together with modeling of complex systems ultimately will allow an understanding of multi-region brain activation, or the way in which various parts of the brain participate in complex tasks such as object recognition, language comprehension, and language formulation. A better understanding of these neural processes in healthy children and adults is essential for the effective study of cognitive disorders. These methods also are the means by which scientists can study the neural reorganization that occurs following brain injury or that results from various kinds of treatment. The use of functional imaging and neural modeling to study the effects of clinical treatment has the potential to improve methods of treatment and to point to new discoveries of brain functions underlying normal and impaired cognitive processes. For example, imaging of the brain has revealed structural and functional differences in the brains of individuals with autism during language processing tasks that are related to the language impairment that is associated with autism. These techniques also have been used to study changes in the brain following therapy for aphasia and stuttering, and they can be applied to many other groups, including individuals with dementia (most of whom have significant language impairment).

Strategic Plan Research Agenda -- Priority Area III

Research is needed to determine perceptual and cognitive processing (how individuals learn to communicate) in normal and disordered communication in the following areas:

- ♦ Use imaging and multi-electrode, multi-unit recording methods such as positron emission tomography (PET), functional magnetic resonance imaging (fMRI), and electroencephalography (EEG) in animal models and humans to dissect the pathways, and define the location and sequence of neuronal activity essential for peripheral and central processing of sensory input. Identify and define abnormalities of neural pathways and spatiotemporal neuronal activity patterns associated with disordered communication.
- ♦ Develop quantitative methods to analyze sensory, sensorimotor, and cognitive processing in humans, in particular, those processes not readily studied in animal models.
- ♦ Investigate the perceptual and cognitive consequences of disordered communication and measure changes that result from treatment, including how the development of language is affected by variations in the quality, quantity, and timing of linguistic input.
- ♦ Investigate how structural and functional differences occur in and affect individuals with communication disorders.
- ♦ Combine cellular, molecular, and physiologic approaches with behavioral analyses in basic science and clinical studies to understand normal mechanisms of sensory processing, cognition and perception.

IV. Develop and Improve Devices, Pharmacologic Agents, and Strategies for Habilitation, Rehabilitation, and Prevention of Human Communication Disorders

It is central to the mission of the NIDCD to enable individuals who have, or who are at risk of having, communication disorders to maximize their quality of life. Therefore, research on intervention for, and prevention of, communication disorders is another research priority for the NIDCD.

Improving, Restoring, or Preventing Communication Disorders

As described in the previous sections, NIDCD-supported scientists have made great progress in recent years toward the goal of understanding human communication and its disorders. These advances were possible because of unprecedented breakthroughs in genetics, as well as other basic sciences and technologies such as microelectronics. As more is learned about the function of the brain and other organs important for communication, this progress will continue as more genes associated with specific communication disorders are identified and their functions are revealed.

Clinical research uses this new knowledge to study human behavior and disease. For example, hearing screening programs around the country are beginning to identify infants and young children who have significant hearing loss. The technology for screening newborns was developed as a result of basic laboratory studies that measured electrical signals from auditory centers in the brain (auditory brainstem response) and sounds generated by the inner ear (otoacoustic emissions). Rigorous clinical trials should be performed to determine the most effective treatments (including hearing aids and cochlear and brainstem implants) and the most effective education programs for hearing-impaired infants, and at what age the treatment should begin in order to achieve maximum success in language development.

Clinical research is also needed to describe the evolution of human communication over an individual's life span, such as hearing, balance, odor detection, or speech abilities. These differences may then be related to an underlying gene or genes, which in turn may help identify, at earlier ages, individuals with a greater risk for developing problems. Once this information is obtained, clinical trials are needed to find safe and effective ways to treat specific communication disorders. These trials may evaluate behavioral interventions, medications, or other therapies. Several examples include laser therapy to treat cancer on the vocal folds, electrical stimulation and medications to treat tinnitus, and physical therapy involving special positioning of the head for loss of balance (positional vertigo).

The NIDCD is committed to conducting and supporting research to develop devices or interventions that improve or restore communication abilities, or prevent communication disorders. For example:

- ♦ Cochlear implants have benefited many children who were born deaf as well as individuals who became deaf later in life. According to the Food and Drug

Administration's 2005 data, more than 96,000 people worldwide have received cochlear implants. In the United States, roughly 22,000 adults and nearly 15,000 children have cochlear implants. Most adults who received implants have benefited greatly and many are even able to communicate effectively by telephone after an extensive training period. Continued research on cochlear implants and sound processing is expected to improve communication of implant users and to improve our understanding of the auditory system. Methods should be developed to assess user performance in order to provide future cochlear implant recipients with more effective implants. Further research is needed to determine whether deaf children will benefit from having implants in both ears. Research is also needed to determine how linguistic input can best help young implant users learn language, to determine what type of input, how much, and during what developmental stage instruction in language is most effective.

- ♦ Although hearing aid technology has advanced rapidly over the past few decades, hearing aids still do not work well where sound comes from more than one source. They are also not particularly effective when a listener tries to pay attention to a single speaker among many competing speakers or in the presence of loud background noise. To meet these needs, improvements continue to be made in developing directional and other hearing aids that will help users understand speech from specific sources within a noisy environment.
- ♦ Use of combinations of a hearing aid and short electrodes for cochlear implants have proven to be effective in allowing individuals with hearing losses in the high frequencies to regain hearing. More work needs to be done to determine which individuals receive and which devices yield the most benefit and when the combination devices should be recommended.
- ♦ Oral communication is often difficult for individuals with severe speech impairments caused by muscular dysfunctions (dysarthria). These impairments are associated with a variety of causes, including trauma and neurodegenerative diseases. However, much progress has been made in developing augmentative or assistive communication devices that help individuals with dysarthria to express themselves. Present research is evaluating whether a low-cost, laser-activated keyboard would enable users to access personal computers. If so, individuals with speech or language disorders could use personal computer programs and speech synthesizers to increase their communication capabilities. The development of automatic speech recognition systems tailored to the speech patterns of individuals with dysarthria may permit computer-based communication using computer-generated speech or text.
- ♦ Advances in basic science research and in bioengineering continue to contribute to: (1) the development of the electro-larynx, which partially restores voice after the larynx is removed; (2) digital programmable hearing aids that fit inside the ear canal; (3) cochlear and brainstem implants, which improve the communication ability of adults and children with profound hearing loss; and (4) video-game-like computer programs that treat disorders associated with childhood language and learning disabilities.

- ♦ By using biological principles of odor recognition, it should be possible to design improved biosensors (electronic noses) that could be used in place of humans and animals, as well as existing biosensors dedicated to detecting specific substances, to detect and discriminate complex chemical signatures of importance to biomedicine, biodefense, and biosafety.
- ♦ Information from epidemiological, biological, and behavioral studies can be used to develop recommendations to prevent communication disorders or to minimize their effects. Advances in knowledge in several areas create new opportunities to identify strategies for prevention.

Important advances have been made in the development of assistive devices, drugs, and other therapeutic interventions, and there is excellent potential for further developments. However, it is important that the basic developmental work be accompanied by clinical research to ensure that these interventions are safe, efficacious, and used to maximum benefit. The ultimate utility and success of current and future devices or other interventions can only be determined by clinical research studies that include the participation of users of the devices or interventions.

Translational research must be promoted to ensure the timely and effective translation of basic research to clinical research. Translation from animal to human, laboratory to bedside, and ultimately from bedside to widespread clinical practice is essential for new knowledge to result in maximum benefit for individuals with communication disorders.

Strategic Plan Research Agenda -- Priority Area IV

Research is needed to improve the quality of life for individuals with communication disorders through assistive devices, drugs, and other therapeutic interventions in the following areas:

- ♦ Capitalize on emerging technologies to design and improve devices that enhance communication, including short electrodes for cochlear implants, brainstem implants, and drug-delivery devices.
- ♦ Use clinical trials and other clinical studies to evaluate the safety and efficacy of newly developed devices, drugs, and other therapies for individuals with communication disorders. These studies should also be used to develop and assess medical and behavioral interventions for infants and children who have a communication disorder.
- ♦ Make early diagnosis and early prevention of communication disorders easier by developing and refining diagnostic criteria and improving diagnostic technology.
- ♦ Screen FDA-approved drugs to determine any beneficial or harmful effect on individuals with communication disorders.
- ♦ Develop cost-effective techniques to assess the variety in patterns of communication, including languages and dialects, currently used in the United States, taking into account the needs of all cultural and ethnic groups.

- ♦ Develop engineered reconstructive tissues that could be used to restore function in individuals who have suffered structural loss through disease or trauma.
- ♦ Integrate information from epidemiological studies with knowledge gained from biological and behavioral research to develop strategies for prevention of communication disorders. Early identification of children at risk for communication disorders is a first step in the prevention of many disorders. Efforts in early identification must be coupled with research to determine the most effective ways to prevent a disorder or to minimize its effects.
- ♦ Identify environmental exposures that contribute to communication disorders, and determine ways of preventing or eliminating these exposures and reducing their harmful effects.

SUMMARY

Disorders of human communication, which when broadly defined include disorders that affect hearing, balance, smell, taste, voice, speech, or language affect millions of Americans. Fortunately, over the past few decades, research has greatly advanced the understanding of human communication and communication disorders. There is a greater understanding of how information is received and interpreted in the brain and how an individual's communication abilities can be compromised by factors such as infection, loud noise, and genetic abnormalities. In addition, many new technologies have been developed to improve or restore communication abilities.

Extraordinary research opportunities led to scientific breakthroughs in the study of genes, proteins, stem cells, and molecular processes that directly affect the understanding of communication disorders. These advances have been accompanied by substantial progress in behavioral studies that increase the understanding of communication processes in health and disease. New imaging techniques, electronic devices, computer databases, animal models, and clinical trials have enhanced our ability to understand, prevent, diagnose, and treat disorders of human communication.

The NIDCD is committed to further advancing the science of human communication and its associated disorders. NIDCD-supported research has been essential to many of these advances but many opportunities remain. The strategic priorities outlined in this plan provide a blueprint for future scientific initiatives and investigator-initiated research aimed at improving the quality of life for individuals facing the daily challenges of living with communication disorders.